

# WILMS TUMOUR WITH INTRACAVAL AND INTRACARDIAC EXTENSION A TRIUMPH IN STAGED SURGICAL PROCEDURE

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# CONFLICT OF INTEREST

- None to declare



# BACKGROUND



- Staged Surgical Procedure (SSP) for management of Wilms Tumour with intracardiac and intracaval extension not commonly reported.
- We describe a case that was successfully managed using SSP approach

# CLINICAL CASE

## • HISTORY

- 11 month old presented with 3/52 history of abdominal distension
- Progressively worsening
- Ultrasound at private hospital revealed a **left sided abdominal mass**.
- Patient transferred to our centre for further management

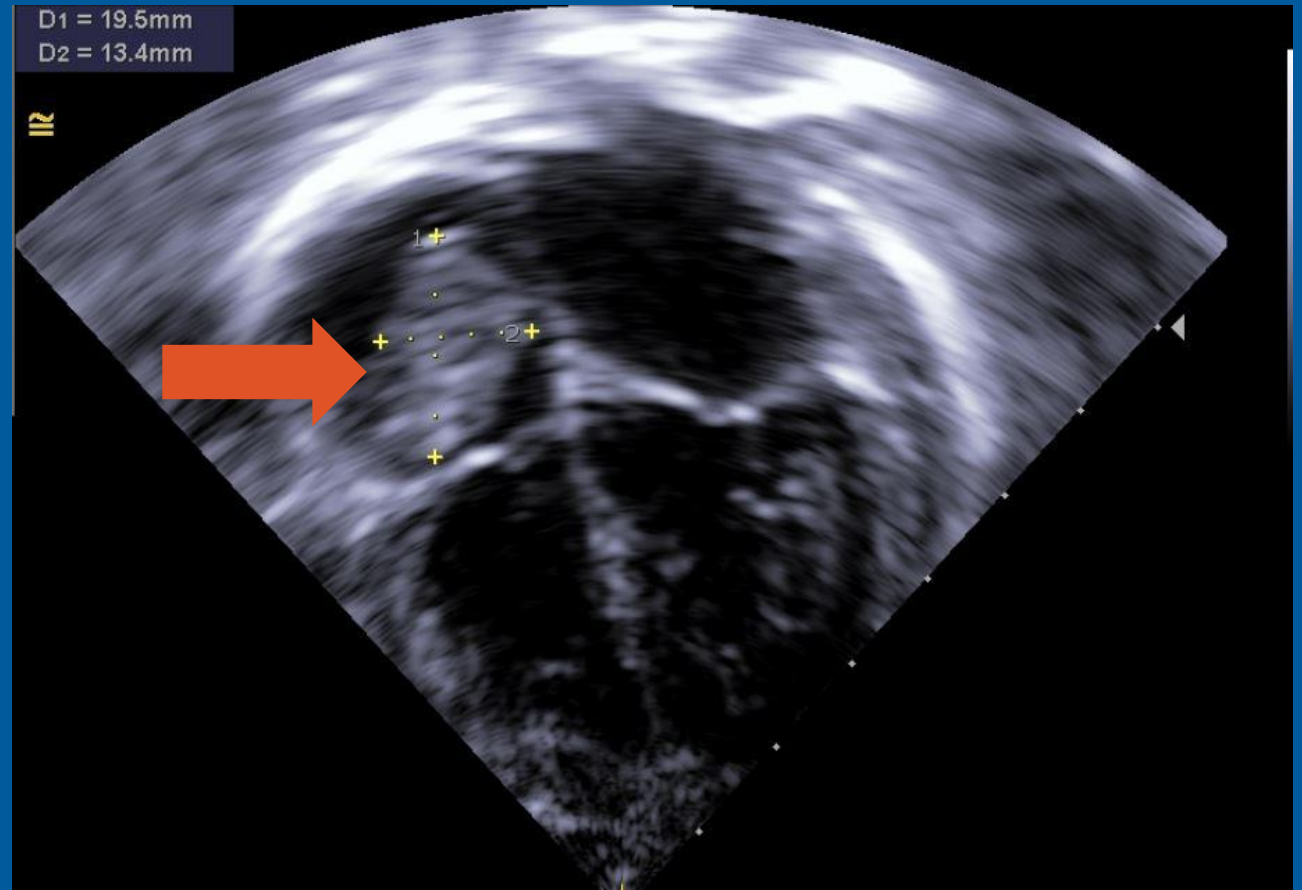
## • EXAMINATION

- Large non tender mass extending from left hypochondrium to left iliac fossa
- No palpable lymphnodes
- LDH 2100

## • USS GUIDED BIOPSY

- Triphasic pattern of tumour cell
- Immunohistochemical staining positive for WT1

# DIAGNOSTIC IMAGING



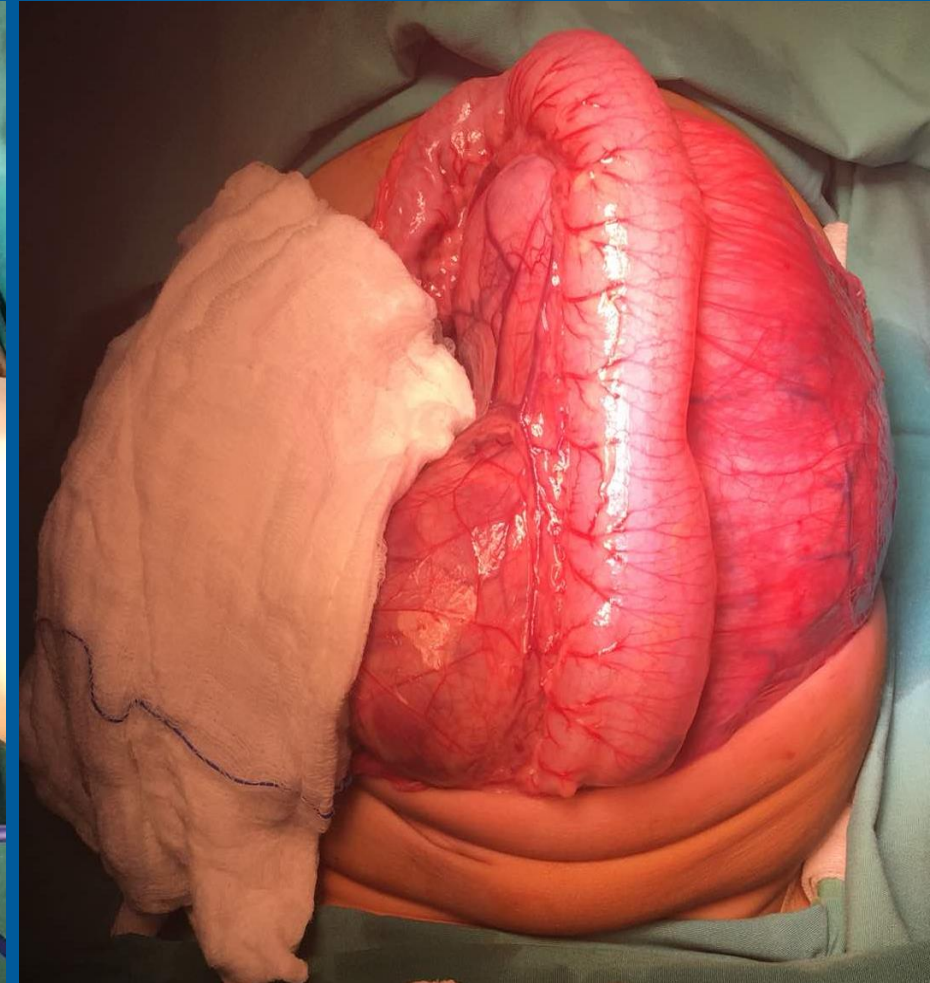
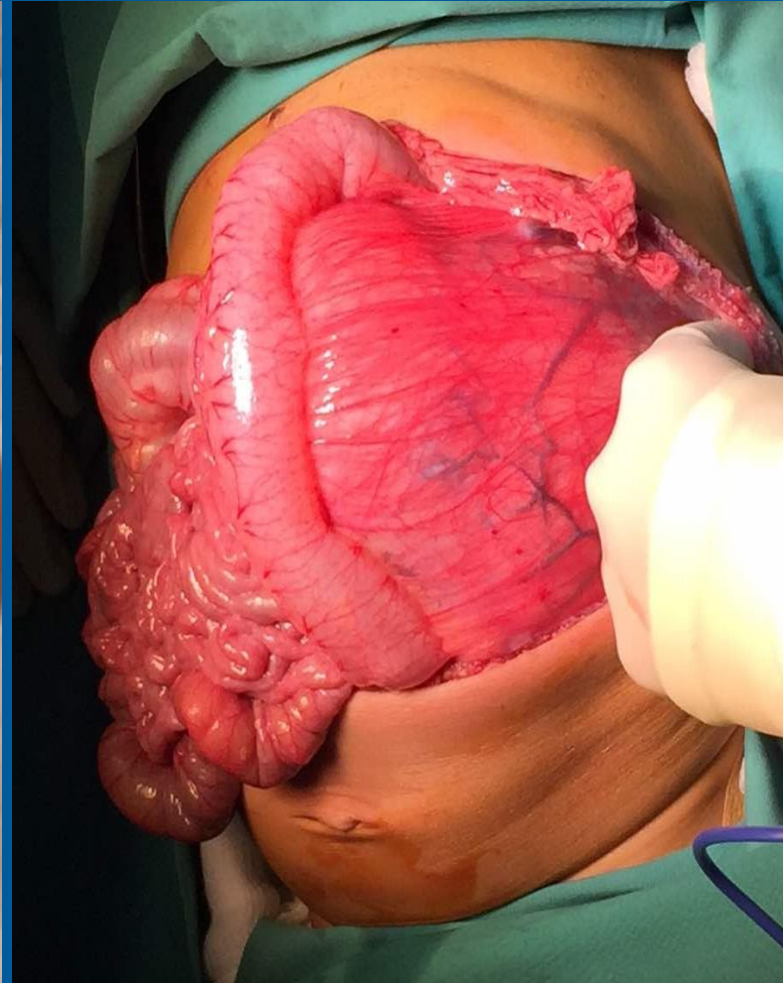
# MANAGEMENT

- **4 cycles** of adjuvant chemotherapy
- **CT reassessment** showed tumour **increasing in size**
- **Repeat ECHO** still shows **thrombus** extending from IVC to right atrium
- Discussion at **Joint MDT** involving 2 different centres - decision for **staged approach** difficulty in arranging joint procedure
- Further cycle of chemotherapy whilst awaiting surgical planning (Cyclophosphamide and doxorubicin)
- Followed by staged procedure: 26/6/2018 + 12/7/2018

# CLINICAL CASE

- **SURGICAL INTERVENTION – 2STAGES**
- **1<sup>st</sup> Stage: Sternotomy and excision of right atrial tumour (26/6/2018)**
  - Intraoperative findings : firm smooth surfaced tumour non adherent to surrounding structures
- **2<sup>nd</sup> Stage: Laparotomy, left nephroureterectomy and removal of tumour in IVC(12/7/2018)**

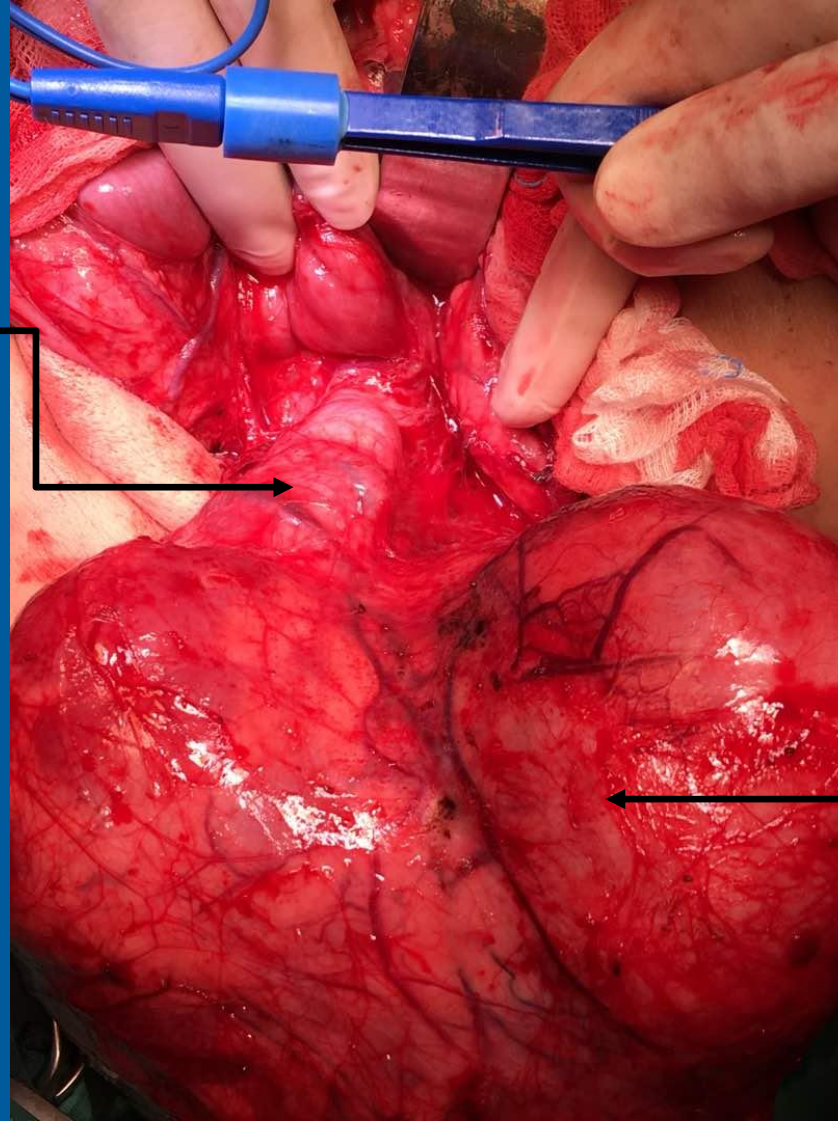
# LAPAROTOMY, LEFT NEPHROURETERECTOMY AND REMOVAL OF TUMOUR IN IVC





# LAPAROTOMY, LEFT NEPHRORECTOMY AND REMOVAL OF TUMOUR IN IVC

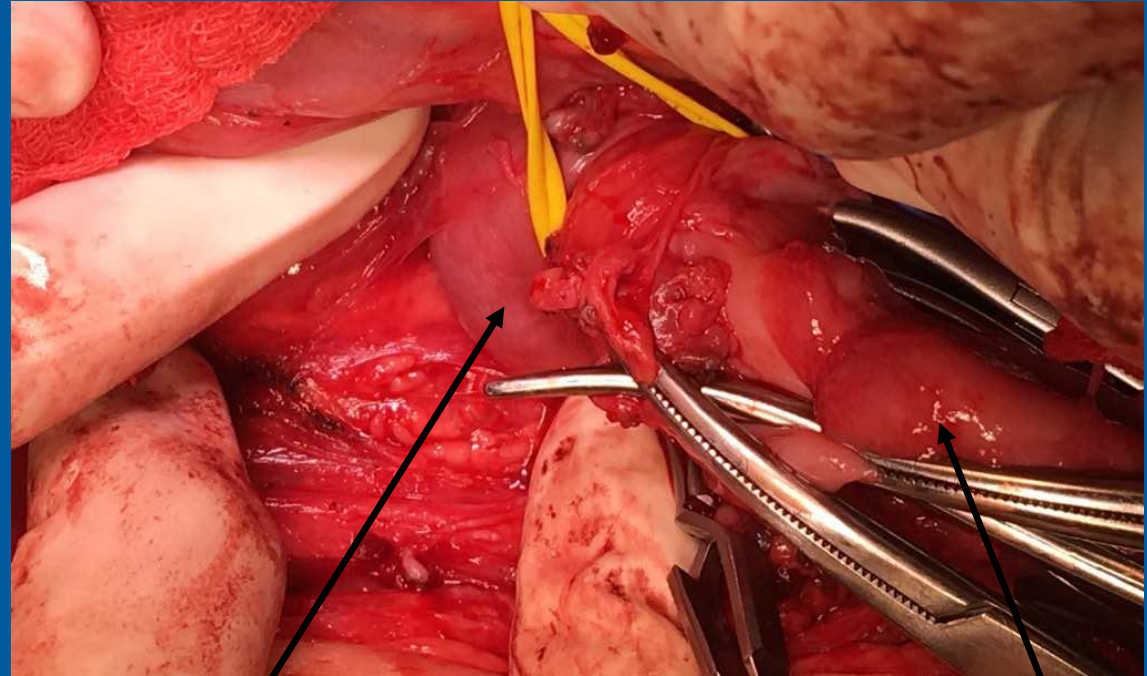
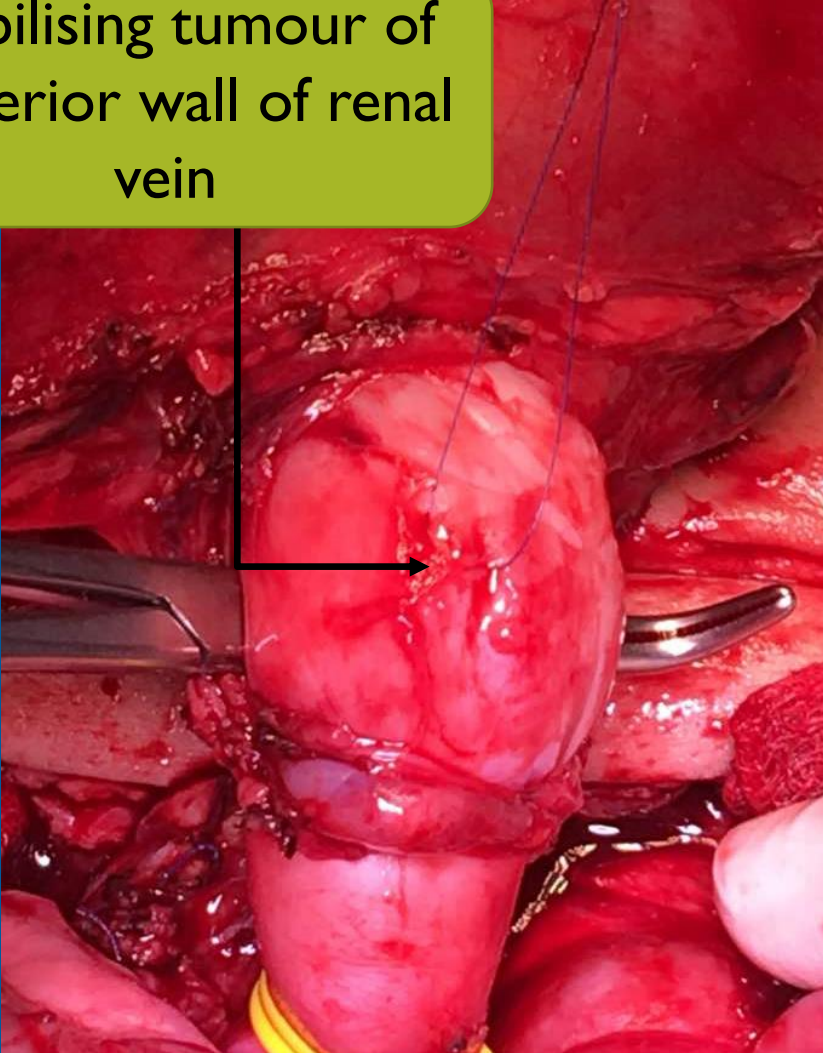
Engorged Left Renal  
vein occluded by  
thrombus



Left Wilms Tumour

# LAPAROTOMY, LEFT NEPHRORECTOMY AND REMOVAL OF TUMOUR IN IVC

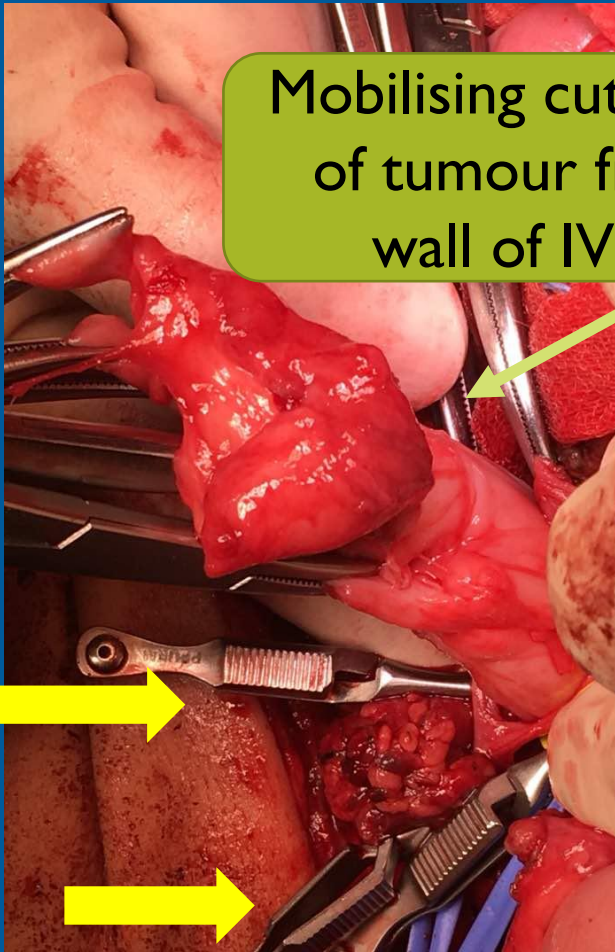
Mobilising tumour of  
posterior wall of renal  
vein



Normal portion of  
infrarenal IVC

Mobilising tumour off  
wall of IVC

Mobilising cut edge of tumour from wall of IVC



Smooth and rounded appearance of superior-most aspect of tumour extending into IVC

BEFORE



AFTER



BEFORE



AFTER



BEFORE



AFTER



# LITERATURE REVIEW

- **I. Wilms Tumour with Intravascular Extension: A Review Article**

- S. McMahon & R. Carachi *J of Indian Assoc Pediatr Surg.* 2014 Oct- Dec;19(4):195-200
- Intracaval Extension 4-8% of all Wilm's Tumour
- Intratrial Extension 1-3% of all Wilm's Tumour
- Management Strategies vary
- Multidisciplinary approach is optimal
- Neoadjuvant chemotherapy advocated
- Surgical resection should be planned in accordance with stage
- Higher rate of complication
- Outcome related to histological subtype

# CONCLUSION

- Remarkable experience for all members of the multidisciplinary team
- Able to deliver good outcome despite challenging circumstances
- Highlights importance of good teamworking
- Surgical Staged Procedure (SSP) can be considered as an option in managing Wilm's Tumour in resource limited settings and should be supported by good multidisciplinary networking.



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*Dayang Aziz*  
#PHOTOGRAPHY

# HISTOLOGY

- **Ultrasound guided biopsy** undertaken:
- **Triphasic pattern** of tumour cell
- **Blastemal component:** undifferentiated small cells hyperchromatic nuclei with scanty cytoplasm
- Loose stroma: **mesenchymal component** consisting of spindle cells with mild pleomorphic nuclei.
- **Epithelial component:** forming tubular/glandular structure
- Immunohistochemical assay : **blastemal cells Positive for WFI**
- Consistent with Wilms Tumour